POSTERIOR POLYMORPHOUS DYSTROPHY OF THE CORNEA*†

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THIS congenital lesion affecting the endothelium and Descemet's membrane is characterized by depressions and bullae on the posterior surface of both corneae usually without stromal involvement. It is not progressive and in the majority of cases causes only slight impairment of vision.

Koepe (1916) described six cases of corneal dystrophy, probably congenital, in which the posterior surfaces of both corneae were marked with small depressions shaped like the segment of a sphere and varying in size. Bullae were not described although these were a feature of cases reported later. Triebenstein (1925) reported three cases, described bullae on the posterior surface of the cornea, and distinguished the condition from Fuchs's endothelial dystrophy and Hassall-Henle bodies. An affected father and son were reported by Freudenthal (1932). Theodore (1939) reported three cases in three generations of the same family, and stated that the condition was separate from Fuchs's dystrophy because it was congenital and not progressive. The youngest of his patients had been seen by an ophthalmologist at the age of 3 months, when the corneae were reported to be affected.

Schlichting (1941) published an extensive review of the previous literature relating to endothelial changes of the cornea, defined posterior polymorphous dystrophy as a separate entity from Fuchs's endothelial dystrophy, Hassall-Henle bodies, and localized bullae of the endothelium. He also described two further cases in a father and daughter. Two affected families were annotated by Cuntz-Schüssler (1947) in one of which three generations were involved; some of her cases were unusual in showing stromal and epithelial changes in addition to the characteristic lesions on the posterior surface. Forni (1951) reported an affected father and daughter, and McGee and Falls (1953) a father and son and an isolated case in which there was a suspicion of parental consanguinity. Snell and Irwin (1958) described a family with seven affected members and Pietruschka (1960) four members of another family in which the condition was associated with glaucoma, vitiligo, otosclerosis, and hypochlorhydria. The corneal findings here were not quite typical, but were classified by François (1966) as posterior polymorphous dystrophy. Other cases have been reported by Kwedar (1961: father and son), and by Hugonniér, Etienne, Bonnet-Géhin, and Laurent (1963: father and two children, and an isolated case with late infantile glaucoma). We do not consider that the cases reported by Bergman (1964) represent true posterior polymorphous dystrophy, as only one eye was affected and only a small area of endothelium was involved.

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Excellent reviews of this condition have been published by Franceschetti, Klein, Forni, and Babel (1951), Duke-Elder (1965), and François (1966).

Case Report

A 6-year-old boy was seen by one of us (H.M.) at a school clinic where he had been referred because of poor visual acuity.

Examination.—He had myopic astigmatism (Right eye -3.75 D sph., -2 D cyl., axis 15° ; left eye -2.5 D sph., -3 D cyl., axis 165°). With spectacle correction the visual acuity was improved to only 6/9 in each eye. On ophthalmoscopy the image of the fundus was seen to be distorted, and this was found to be due to fine irregularities on the posterior corneal surface. Later slit-lamp examination showed that all areas of the posterior surface of both corneae were covered with groups of vesicles and depressions, none larger than 0.5 mm. A fine grey opacity in Descemet's membrane surrounded each group. The endothelium between the groups of vesicles and depressions appeared normal as did the stroma and epithelium (Fig. 1). The corneal sensation appeared normal.

FIG. 1.—Sketch of parallelopiped of cornea seen in the slit-lamp beam.

FIG. 2.—Family tree of case described.

Family Study.—Fig. 2 shows that no affected relatives could be discovered, but the dearth of surviving maternal relatives makes elucidation of the mode of inheritance impossible. There was no history of parental consanguinity.

Discussion

Most of the cases previously reported have shown a dominant mode of inheritance (Freudenthal, 1932; Theodore, 1939; Schlichting, 1941; Cuntz-Schüssler, 1947; Forni, 1951; McGee and Falls, 1953; Snell and Irwin, 1958; Pietruschka, 1960; Kwedar, 1961;

Hugonnier and others, 1963). Neither Koeppe (1916) nor Triebenstein (1925) reported the family histories of their cases.

A recessive mode of inheritance is perhaps demonstrated by the isolated case described by McGee and Falls (1953), where there was a possibility that the parents were related, and by the two sisters reported by Soukup (1964).

Summary

An isolated case of posterior polymorphous corneal dystrophy is described. We believe this to be the first case reported from the United Kingdom.

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ADDENDUM

Since this paper was submitted, Morgan and Patterson (1967) have reported the pathology of a case of posterior polymorphous dystrophy.